

Spontaneous carotid-cavernous fistula: a case report

Spontaneous carotid-cavernous fistula

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Abstract

Aim: Carotid-cavernous fistula is an arteriovenous shunt that develops in the cavernous sinus. It can occur either following a head injury or spontaneously. Posttraumatic carotid-cavernous fistulae are relatively easy to be diagnosed from history and clinical presentation. However, spontaneous carotid-cavernous fistulae which present without a history of trauma or a vascular disorder may be rather difficult to identify. **Results:** We present a 54-year-old lady with the initial clinical diagnosis of an orbital pseudotumor and was later diagnosed as having carotid-cavernous fistula on magnetic resonance imaging. **Discussion:** A high index of clinical suspicion and a thorough radiologic evaluation is needed for accurate diagnosis of spontaneous carotid-cavernous fistulae.

Keywords

Carotid- Cavernous Fistula; Fistula; Magnetic Resonance Imaging

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Introduction

Carotid-cavernous fistula (CCF) is an arteriovenous shunt that develops in the cavernous sinus. Most CCFs are posttraumatic, usually secondary to skull base fractures, and typically present with painful pulsatile exophthalmos, glaucoma, orbital edema, decreasing vision, and headache [1,2]. The diagnosis of post-traumatic CCFs is relatively easy detected from history and clinical presentation. However, spontaneous CCFs which present with relatively mild ocular manifestations, especially the ones occurring in previously healthy individuals, may be rather difficult to identify. The diagnosis of spontaneous CCFs is based on a high index of clinical suspicion and a critical radiologic evaluation [2]. Herein we present a 54-year-old lady with the initial clinical diagnosis of an orbital pseudotumor and was later diagnosed as having CCF on magnetic resonance imaging (MRI).

Case Report

A previously healthy 54-year-old woman who presented with headache, severe proptosis, dimness of vision and pain of the right eye for the last six weeks was referred to the Department of Radiology for a brain MRI. The initial clinical diagnosis was an orbital pseudotumor. On T1 and T2-weighted images, there was a severe proptosis of the right eye with enlarged extraocular muscles. The right ophthalmic vein was dilated (Figure 1,2). Contrast-enhanced 3D TOF magnetic resonance angiography (MRA) depicted remarkable dilatation of the right cavernous sinus in which there was early enhancement due to high-flow (Figure 3). The right ophthalmic vein was prominently larger than the left one (Figure 4). Based on the typical imaging findings, the patient was diagnosed as having CCF.

Discussion

CCFs are abnormal communications that develop between the carotid arterial system and cavernous venous sinus. They are divided into two categories: direct and indirect. Direct CCFs, which account for approximately 75% of all CCFs, result from rupture of the internal carotid artery directly into the cavernous sinus. Most direct CCFs occur following a head injury, usually

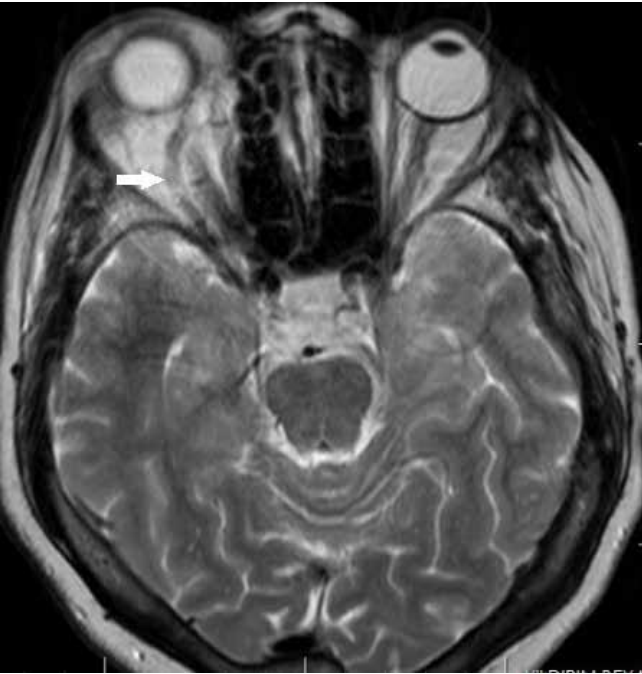


Figure 1. Axial T2-weighted image demonstrates a striking proptosis and an enlarged superior ophthalmic vein (arrow).

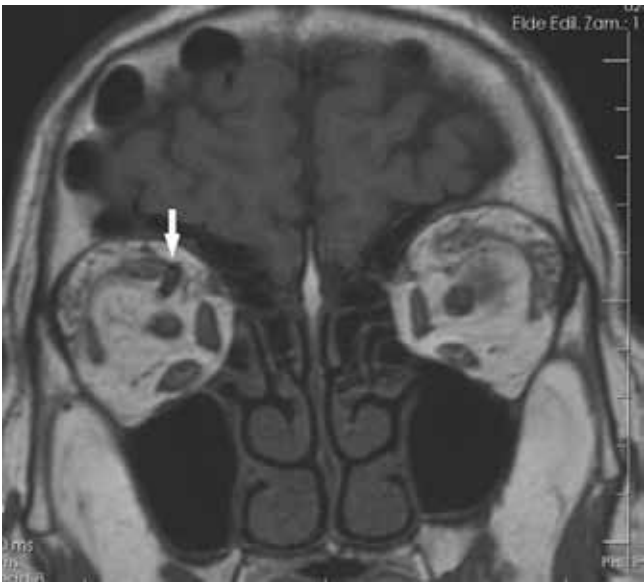


Figure 2. Coronal T1-weighted image shows the prominent enlargement of the right superior ophthalmic vein (arrow). Note enlarged right extraocular muscles compared to their left counterparts.

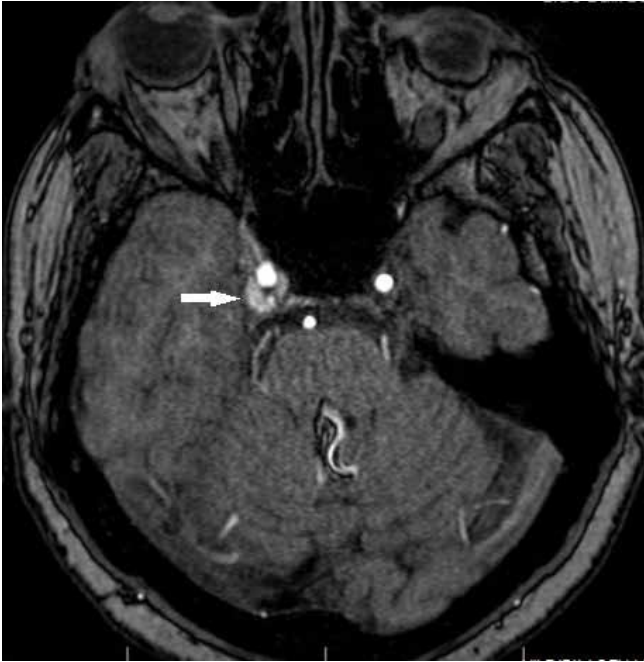


Figure 3. Contrast-enhanced, 3D TOF magnetic resonance angiography demonstrates a dilated right cavernous sinus in which there was an early enhancement due to high-flow (arrow).

after a central skull base fracture. Indirect CCFs, or spontaneous CCFs, are nontraumatic lesions which occur secondary to spontaneous rupture of one of the thin-walled dural arteries passing through the sinus. They occur spontaneously, especially in middle-aged to elderly women, and may be associated with atherosclerosis, systemic hypertension, collagen vascular disease, pregnancy, connective tissue disorders, and minor trauma. Spontaneous CCFs account for approximately 25% of all CCFs [1,2]. The diagnosis of CCF in patients without a history of trauma or a vascular disorder is somewhat challenging as was the case in our patient. They may be mistaken for chronic conjunctivitis, orbital pseudotumor, orbital cellulitis or thyroid eye disease[2].

Computerized axial tomography (CT) and MRI are the preferred radiologic modalities for the diagnosis of CCF. Compared with

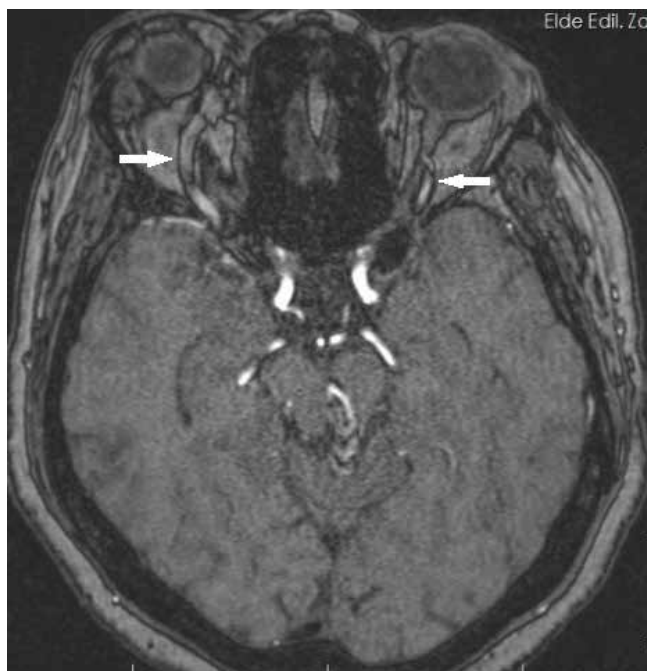


Figure 4. On contrast-enhanced 3D TOF magnetic resonance angiography, the right superior ophthalmic vein is more than 4 times the size of the left superior ophthalmic vein (arrows).

angiography, CT and MRI have a much lower incidence of complications, furthermore, they depict peripheral pathologies associated with CCFs. Digital subtraction angiography is used to confirm CT or MRI findings prior to the treatment [1,3,4]. On CT or MRI, the characteristic findings of a CCF are proptosis of the affected eye and enlargement of the ipsilateral superior ophthalmic vein, cavernous sinus, and extraocular muscles. If the superior ophthalmic vein appears to be either asymmetric or larger than 4 mm in diameter, CCF is suggested. MRI is better in evaluating venous distension and the increased flow to cavernous sinus compared to CT. When it is used in conjunction with MRA, even better diagnostic capability is achieved [1,2]. Our patient clearly showed the imaging characteristics of CCF on both conventional MRI sequences and MRA, and the diagnosis was established without a need for DSA.

Endovascular embolization is an established modality in the management of CCFs. All cases of direct CCF are treated interventionally, whereas only selected symptomatic cases of indirect variety need to be treated [5,6].

Conclusion

Compared to direct CCFs, spontaneous CCFs are rare and may be difficult to diagnose. A high index of clinical suspicion and a thorough radiologic evaluation is needed for an accurate diagnosis.

Scientific Responsibility Statement

The authors declare that they are responsible for the article's scientific content including study design, data collection, analysis and interpretation, writing, some of the main line, or all of the preparation and scientific review of the contents and approval of the final version of the article.

Animal and human rights statement

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. No animal or human studies were carried out by the authors for this article.

Conflict of interest

None of the authors received any type of financial support that could be considered potential conflict of interest regarding the manuscript or its submission.

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